

SEER SINC

Finalized October 2010

Question: [20100033](#)

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Question

Heme & Lymphoid Neoplasms--Histology: What histology is to be coded for a case that is described as follicular lymphoma, grade 3a/3 (9698/3), with focal areas of diffuse large B cell lymphoma (9680/3) (approximately 10%)? See discussion.

Discussion

Per rule PH16, Code the primary site to the site of origin (lymph node region(s), tissue, or organ) and code the histology diffuse large B-cell lymphoma (DLBCL)(9680/3) when DLBCL (9680/3) and follicular lymphoma (9690/3) are present in the same lymph node(s), tissue, or organ.

Does the term "focal" have the same significance (ie, ignore) in Heme cases as it does for solid tumors? Also, does the follicular lymphoma, have to be NOS (9690/3) or does PH16 include all grades of follicular lymphoma (9695/3, 9691/3, 9698/3)?

Answer

Code the histology 9680/3 using rule PH16. Follicular lymphoma (FL) includes FL, NOS, FL grade 1, FL grade 2 and FL grade 3. Focal, foci, and focus are not used in the hematopoietic rules, meaning that you do NOT ignore histology terms described as focal, foci, or focus.

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10/04/10

Question: [20100034](#)

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Question

MP/H Rules/Multiple primaries--Esophagus: In light of the fact that tumors of the GE junction are now included with tumors of the esophagus in AJCC 7th Edition, will the MP/H rules also be adjusted to reflect that change? Should a patient with separate tumors in the esophagus and in the GE junction be accessioned for one or two primaries? See discussion.

Discussion

Per notes included in CSv2, the cardia/EGJ, and the proximal 5cm of the fundus and body of the stomach (C16.0-C16.2) have been moved from the Stomach chapter and added to Esophagus effective with AJCC TNM 7th Edition. A new schema EG Junction was created in CSv2 to accommodate this change. Tumors arising at the EGJ, or arising in the stomach within 5 cm of the EGJ and crossing the EGJ are staged using the schema for EG Junction. MP/H Rule M11 states that tumors with ICD-O-3 topography codes that are different at the second (Cxxx) and/or third characters (Cxxx) are multiple primaries. Example: Patient with two nodules of adenocarcinoma, one at the GE junction (C160), the second in Barretts esophagus, distal esophagus (C155). How many primaries should be accessioned for this case? Should the two tumors be counted as a single primary (change in MP/H rules) because they are grouped together in the same stage grouping? Or did CSv2 create a separate schema for Esophagus GE Junction to accommodate multiple primary cases in which separate tumors arise in the esophagus and in the cardia/GE junction?

Answer

Use the multiple primary rules to determine the number of primaries. Use staging resources for staging. Abstract two primaries for the case example using rule M11.

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Question

MP/H Rules/Multiple primaries—Colon: How many primaries are to be counted for a patient with two colon carcinomas? There is no documentation that either tumor arose in a polyp and there is no statement indicating the presence of adenomatous polyposis coli, however, path documents the presence of over 200 polyps in the resected specimen. See discussion.

Discussion

The first MP/H rule that applies for our example case is M4. Should rule M3 be expanded to include a statement about > 100 polyps? If we follow rule M4, case would be counted as two primaries. We would never get to rule H17 (code 8220 [adenoca in adenomatous polyposis coli] when there are > 100 polyps identified in the specimen) because H17 applies to multiple tumors abstracted as a single primary. Example: Total colectomy: Distal tumor: - ulcerating moderately differentiated colonic adenocarcinoma, 3.2 cm in greatest dimension. - tumor invades through the muscularis propria into the subserosa (pt3). Proximal tumor: exophytic moderately differentiated colonic adenocarcinoma, 2.9 cm in greatest dimension. Tumor invades submucosa (pt1). Multiple tubular adenomas present throughout the colon, approximate count greater than 200.

Answer

Use rule M3 for this case and abstract as a single primary. The case information makes it clear that this is adenomatous polyposis coli. Clarification will be added to rule M3 in the next revision of the rules.

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Question

Behavior--Lung: How is this field coded for a "bronchioloalveolar carcinoma of the lung?" See discussion.

Discussion

In ICD-O-3, bronciolo-alveolar adenocarcinoma is described only by behavior code 3 (invasive). However, pathologists appear to use the term bronchoalveolar to describe an in situ pattern of growth exhibited by adenocarcinoma. Example 1: Left lower lobe, partial resection shows bronchioloalveolar carcinoma with focal areas of fibrosis (see comment). Comment: Although the possibility that these areas represent stromal invasion can not be excluded, we favor the interpretation that these areas do not represent true invasion. Synoptic summary: Minimal pathologic stage: Local Extent. Example 2: Lung tumor described as adenocarcinoma, predominantly bronchoalveolar pattern.

For most sites, pattern is used only for in situ cancer and is not a specific term used for invasive tumors. Is the use of the term "pattern" in this situation indicative of in situ tumor?

Answer

Code the behavior as indicated by the pathology report. If the pathologist states that bronchioloalveolar carcinoma is in situ, apply the ICD-O-3 matrix concept and assign 8250/2. Otherwise, code 8250/3. Do not use the term "pattern" to infer in situ behavior.

Code behavior /3 for both examples based on information provided.

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Question

Multiple primaries/Histology--Heme & Lymphoid Neoplasms: How many primaries should be abstracted and what are the histology codes for this case? See discussion.

Discussion

Patient had a diagnosis of essential thrombocythemia (9962/3) in 2002 and was treated with Hydroxyurea. In 2010, the patient was admitted with severe bone pain and the diagnosis is, "The overall features of the biopsy are consistent with a fibrotic stage of a chronic myeloproliferative disorder. ... The presence of up to 15% CD34+ immature cells seen in the biopsy suggests that the patient is transforming to an acute myeloid leukemia." In addition, cytogenetic studies and molecular testing for JAK2 were ordered. These findings confirmed a myeloproliferative disorder. JAK2 mutation was not detected. The patient died within 2 weeks. Is this a new primary? Is it AML (which requires 20% or more blasts and this is only 15%)? If so, is it 9861/3 or 9895/3? [I think it should not be a new primary because the second diagnosis of AML is not definitively diagnosed.]

Answer

This case is a single primary (essential thrombocythemia in 2002). The 2010 diagnosis is chronic myeloproliferative disorder (9960/3). According to the multiple primaries calculator, essential thrombocythemia and chronic myeloproliferative disorder are the same primary. Acute myeloid leukemia is not reportable in this case because it is preceded by a non-reportable ambiguous term: "Suggests" is not on the list of reportable ambiguous terms. See page 12 of the 2010 Hematopoietic and Lymphoid Neoplasm manual. Note: Ambiguous terminology is used to determine reportability; it is NEVER used to determine histology for heme and lymphoid neoplasms.

In 2010, this patient was in a late stage of ET. When any of the specific MPN neoplasms such as ET are in the late stage of disease, the characteristics of the specific disease (ET) will no longer be detectable. Accordingly for this patient, the diagnostic testing was positive for MPN, unclassifiable. In this case, do not change the diagnosis from the more specific disease (ET) to the NOS (MPN, unclassifiable).

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Question

Heme & Lymphoid Neoplasms: For patients who have a chronic hematologic disease, such as CML, or myeloproliferative disorder such as essential thrombocythemia and are treated with long-term chemotherapy, is their tumor status always going to be "not disease free" or, when the physician documents that they are in clinical remission, does their status change to "NED or disease free?" For some patients, the disease / recurrence status could change frequently as chemo is started and stopped. There seems to be a lot of variation across the US how registrars are coding this and any clarification would be appreciated.

Discussion

Answer

You are correct, the term "disease free" is not used in a standard fashion for hematopoietic and lymphoid neoplasms.

Code as free of disease when physician states NED. For hematopoietic and lymphoid neoplasms, a physician's statement of NED, disease-free, or no evidence of disease at this time, should be recorded as cancer status: disease free. The term "disease free" or NED means that there is no clinical evidence of disease.

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Question

Reportability--Heme & Lymphoid Neoplasms: How should we handle "myelodysplasia?" Is it reportable?

Discussion

Answer

The diagnosis of myelodysplasia itself is not reportable because the term includes both non-malignancies and malignancies. Follow-back to the physician is necessary to determine whether or not this is a malignancy.

The following definition will be added to the glossary in the Hematopoietic and Lymphoid Neoplasm manual. The term "myelodysplasia covers a group of disorders that result in the inability to produce enough healthy mature blood cells. Those

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disorders include: Anemia, leukopenia, thrombocytopenia, MDS, refractory anemia, refractory anemia with excess blasts in transformation, refractory anemia with ring sideroblasts, refractory anemia with excess blasts, chronic myelomonocytic leukemia, acute myeloid leukemia.

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10/08/10

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Question

Primary site--Heme & Lymphoid Neoplasms: What primary site code should I use? Patient was diagnosed with Langerhans cell histiocytosis/eosinophilic granuloma, biopsy of 7th rib on 3/22/10. On 4/13/10 patient had a right external ear canal mass (right temporal bone) with same diagnosis. Is the primary site coded to bone, nos?

Discussion

Answer

Code the primary site to bone, NOS. Langerhans cell histiocytosis can occur as a solitary lesion, multifocal lesions, or multisystem disease. In this case, the patient has multifocal disease of the bone. The abstractor notes in the Hematopoietic DB were used as a reference for this answer.

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Question

Multiple primaries--Heme & Lymphoid Neoplasms: See Discussion for case description. Could you please walk me through the process of determining if this is one or two primaries.

Discussion

Case: CT scan showing retroperitoneal and inguinal adenopathy. Right inguinal lymph node biopsy revealed "malignant lymphoma, peripheral t-cell type, with some features of angioimmunoblastic t-cell lymphoma and follicular t-cell lymphoma." FLOW showed no evidence of B-cell lymphoma and atypical CD3+/CD10+/CD7-/CD4+/CD56+ T cells are detected (19%). Bone marrow biopsy was neg for involvement. Patient was staged as Stage II Peripheral T-Cell lymphoma by the oncologist and started chemotherapy.

Answer

Code the oncologist's clinical diagnosis of peripheral T-cell lymphoma.

Step 1: Search the DB by entering the unique word "peripheral" in the search engine. The first entry in the matched term list is peripheral T-cell lymphoma, NOS. Now you have a PROVISIONAL ICD-O-3 code and diagnosis.

Step 2: Confirm that this diagnosis fits the case you are abstracting. Click the display button on the mid-bottom of the screen. The definition for this neoplasm is "A large group of lymphomas with the addition of "unspecified" to emphasize that these cases do not belong to an better defined entities. Attempts to identify a more specific disease were unsuccessful."

Step 3: Look at the abstractor notes. Click on "Display Abstractor Notes" on the mid-bottom of the screen. Selected parts of the information available are: Patients present with peripheral LN involvement; The diagnosis of PTCL, NOS is made ONLY when other specific entities have been explored.

This fits your case; attempts to find a more specific disease (flow cytometry; BM biopsy) were negative and gave no further information that could be used to assign a more specific classification.

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Question

Reportability: Do I report carcinoid tumor of the appendix when no other explanation is given? I currently have a case where there is perineural invasion present. Is this reportable? Is it reportable if angiolymphatic invasion is present? The word malignant is not used in the pathology report.

Discussion

Answer

Carcinoids of the appendix are reportable when they meet any of the following conditions:

1. The pathologist designates the carcinoid as malignant
2. Regional lymph nodes are positive for MALIGNANT carcinoid (not reportable if LN are reported as benign carcinoid)
3. There are discontinuous metastatic implants or involvement.

Note that the implants/involvement must be designated as malignant. Many benign tumors will spawn implants that are also benign. If implants are benign, this is not a reportable tumor.

Neuroendocrine carcinoma is an NOS term that includes carcinoid tumors. The reportability requirements for CoC, the National Program of Cancer Registries (NPCR) CDC, and SEER National Cancer Institute have not changed. Carcinoids of the appendix are only reportable when they meet the criteria listed above.

The MPH rules are currently under revision and the implementation date is 2012. This issue will be discussed by the GI specialty physicians, the ICD-O-3 editors and the MPH committee. Any change in reportability would be published in those rules and implemented in 2012. In the meantime, follow the reportability rules as published by the organizations to which you report cases.

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Question

Primary site--Heme & Lymphoid Neoplasms: Patient was diagnosed with Myeloid Sarcoma (granulocytic sarcoma) by chest wall bx. Whenever I enter the information for this case using C495 as the site code, I get an edit error that questions this histology for this site. If I choose bone marrow as the site I do not get an edit error. If this is an extramedullary manifestation of acute leukemia and is not in the bone marrow (bone marrow is negative) which site code should I be using? Rule PH15 in the data base states to code site of origin lymph node region tissue or organ. In my case the only involvement is the chest wall. Could you explain site code and rationale?

Discussion

Answer

Unless there are scans showing involvement of LN or tissue other than chest wall, the histology should be coded myeloid sarcoma 9930/3 and the primary site to C493 soft tissue of chest wall. Override the edit.

The steps taken to resolve this issue are:

1. Find the provisional ICD-O-3 histology code and confirm site/histology by searching the Hematopoietic DB for myeloid sarcoma. The DB shows myeloid sarcoma as the first result. There is a "warning note" that says see Module 5 PH15. Write this module and rule number down. Next get the ICD-O-3 code 9930/3. Confirm the site/histology combination by displaying the abstractor notes. The notes say the most frequently affected sites are skin, LN, GI tract, soft tissue, and testis.
2. Reportability Instructions: The proposed ICD-O-3 code fits in the range of reportable neoplasms
3. Multiple primary rules: M2 says a single histology is a single primary
4. PH Rules: Go to Module 5 PH15 as the warning module in the Hemato DB instructed. The rule says code the primary site to the site of origin. Note 1 again reinforces that the common primary sites are skin, LN, GI tract, bone, soft tissue, and testis. Code to the soft tissue of chest wall.

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Question

Primary site--Heme & Lymphoid Neoplasms: Myeloid sarcoma, tumor mass of myeloblasts or immature myeloid cells occurring in a site other than bone marrow.

Edits do not allow other sites for 9930/3, only C42.1. So must we code these to C42.1? See discussion.

Discussion

Scenario: C03.0 upper gum 9930/3 myeloid sarcoma. Cannot code to primary site; histology is excluded.

Scenario: C341 lung 9930/3 myeloid sarcoma. CS exclusion of histology 99303 for lung.

Answer

Over-ride this edit. Myeloid sarcoma does NOT originate in bone marrow. This has been sent to the edits committee with a request that the edit be changed.

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10/26/10